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A case of Urticaria pigmentosa



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A CASE OF URTICARIA PIGMENTOSA.¹

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IN looking over the literature of urticaria pigmentosa the impression was forced upon me that there was in almost all the published cases a marked similarity in the objective phenomena: the subjective symptom of itching was found, it is true, somewhat variable, and in a few cases, as in that about to be reported, practically *nil*. Itching is to be considered as almost a *sine qua non* of urticaria, and this is always of an annoying character. Indeed, in studying the cases of urticaria pigmentosa carefully, I am constrained to confess that while many of the symptoms point to a close resemblance to true urticaria, in other respects they are so different that as yet their identity can by no means be admitted. This impression is further strengthened by the fact that these anomalous cases stand so prominently divided from simple urticaria, that midway cases (if the expression may be allowed) are not encountered—it is either urticaria, with its evanescent and characteristic wheals, etc., or it is urticaria pigmentosa with its usual symptoms. This sharp division is not in accord with our experience in other cutaneous diseases. The following brief notes, therefore, I wish to put on record, so as, perhaps, to add something to the final status of these cases.

The patient, a boy of six years, came under my notice in the early part of last February. He was a blond, of robust physique, and in perfect health. In fact, no departure from health could be elicited other than the skin eruption for which advice was sought. The parents were also free from disease. In earlier life, and also in the summer preceding the patient's birth, the mother had had urticarial attacks of the usual evanescent type, and due to dietetic indiscretions. The boy's sister, the only other child, had always been free from any cutaneous disease.

The eruption began, as the father stated, after two unsuccessful attempts at vaccination, when the boy was eighteen months old. Since that time, a period of four and a half years, the disease had continued uninterruptedly. The eruption appeared without any

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attempt at regularity, new lesions, few or in numbers, making their appearance from time to time. The covered regions of the body were the parts upon which the spots were most abundant. Lately, however, there had been a strong disposition to appear on the face, and it was this disfigurement chiefly that had made the parents solicitous of remedial help. The lesions had always been numerous about the neck, sides of the trunk, and about the genitalia; until recently comparatively few had been on the limbs and face. The eruption has been active ever since its first appearance, the older lesions eventually disappearing without leaving a trace. Itching had never been a troublesome symptom.

At the time of examination the eruption was found to be more or less general, being most abundant on the lateral portions of the chest and abdomen, on the neck, on the under part of the lower jaw, on the forehead and temples, on the back of the neck well up to the post-aural regions, and to a less extent on the arms and legs. On the face proper there were comparatively few. The eruption consisted of pin-head to large pea-sized rounded, some elongate, reddish-yellow, papule-like elevations; many similar elevations surmounted with a small vesicle with a somewhat thick and light-yellowish epidermal covering; flattened, elevated, small and large pea-sized yellowish or salmon-colored lesions; small spots of pale-yellowish pigmentation, with slight, if any, elevation; spots of similar size, of the color approaching the normal skin, with possibly a yellowish tinge, over which the epiderm appeared slightly loose and wrinkled, and in some the follicular outlets seemed enlarged, giving the appearance faintly similar to the slight atrophy which follows lupus erythematosus. Some of the retrogressing lesions, those which were still somewhat elevated, bore a slight resemblance to both lupus deposits and to xanthoma. The most prominent color was yellow, somewhat on the salmon. In the early life of a lesion, however, a reddish tint was probably the more noticeable. The first appearance of a lesion was, in fact, somewhat similar to the wheal of an ordinary urticaria, although smaller and less inflammatory. Such beginning lesions were to a slight degree itchy, but this was evanescent and never marked or persistent. The skin on being rubbed exhibited a slight tendency to become urticarial.

This was, in brief, the extent and character of the eruption when first seen, and it may here be added also that in the examinations made subsequently the same essential phenomena presented. The lesions, no doubt, finally disappeared, probably in the course of months, without leaving a trace; at least this must be inferred from the fact that the disease had already lasted four and a half years, with continuous outbreak of new lesions, and yet with so comparatively few of the atrophic-looking and the freckle-like spots remaining. These last-named would undoubtedly give place to normal skin. The fact of the total disappearance of the lesions the father also attested.

An observation extending over some weeks showed the eruption in its formation and evolution as follows: The lesions began as small rounded, solid elevations, reddish-yellow in color, developing suddenly or in the course of several hours or days. On many of these a more or less perfect attempt at vesiculation occurs. The vesicle is small and the

epidermal covering thick, giving the vesicle a yellowish color, although the contents remain clear and purely serous. The vesicle disappears by absorption. The spots, as a rule, become a trifle larger, after they are apparently fully developed; they flatten out, and then look like flat, pea-sized yellowish or yellow-brown maculo-papules, being a part apparently of the skin itself; suggesting in some respects a slightly elevated hairless pigmented nævus, and in other respects a superficial flattened xanthoma. Still further flattening gradually takes place, and the epidermis covering the lesion assumes a slightly loose or wrinkled appearance, in some with a tendency toward thinning or atrophy, with also an apparent, and probably true, enlargement of the follicular outlets, suggesting, as already remarked, a faint resemblance to the sieve-like atrophy of lupus erythematosus. These characters—the atrophic and wrinkled appearance and the enlargement of the follicular orifices—could only be seen, however, by close inspection, and were not, therefore, in the slightest degree striking or conspicuous. Indeed, unless the skin were carefully examined these later changes would entirely escape observation. Nor were these characters an essential part of each disappearing lesion. Many spots apparently disappeared without this preliminary atrophy taking place. Whether atrophic or otherwise, however, no permanent trace of the eruption was left, the skin finally assuming its natural condition and color. These several stages—the formation, evolution, and disappearance of a lesion—required weeks, and doubtless in some lesions months. As it was, unfortunately, impossible to gain the parents' permission to excise a lesion, no microscopical examination could be made.

A few words as to treatment. In the reported cases of this disease treatment, except the partial relief to the itching by external applications, was practically negative, and the same may be said indeed in regard to the case here given. Treatment is essentially theoretical and experimental, and must necessarily remain so as long as the nature of the disease and its underlying causes are unrecognized. Before coming under my observation this boy had been variously treated, in one instance taking fair doses of arsenic continuously for several months, but without the slightest effect upon the eruption. With the idea of possibly influencing the vaso-motor nerves and indirectly the disease, the fluid extract of ergot was prescribed, at first in ten minim doses t. d., later increased to twenty minims, and the same was continued for five weeks, but the condition was in no way modified or improved, the older lesions gradually changing as formerly and new lesions appearing from day to day and week to week. This was now discontinued, and the phosphates in combination with cod-liver oil given as a nutrient tonic. I may add that there was no other indication for this than that the boy was of the appearance and complexion so common to strumous subjects, who while

presenting a tolerably robust appearance are lacking in nervous tone and muscular vigor. At the end of a month the disease appeared less active, but this was in all probability, however, independent of treatment, as not infrequently before, so the father stated, there had been times when the disease was comparatively quiescent. The same remedies, with the addition of a small tonic dose of arsenic were continued, and so far as I have been able to learn this treatment was followed for several weeks longer, but as it was without further appreciable effect, was discontinued, and other advice sought.



